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Case Report

Bilateral Mooren's ulcer – Customised corneal graft with additional amniotic membrane graft

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Abstract

Mooren's ulcer (MU) is a rare and painful peripheral corneal ulceration which occurs in the absence of any associated scleritis, and any detectable systemic disease. A 60-year-old male patient was referred to us with bilateral peripheral corneal ulceration. Best corrected visual acuity (BCVA) in both eyes was counting finger at one metre. The right eye showed a 180° thinning with perforation at 8 o'clock position. The left eye showed a 360° thinning with central contact lens type cornea. After complete blood analysis we started the patient on cyclophosphamide orally along with high doses of oral steroids. A crescentic excision of the thinned cornea and crescentic customised corneal graft with additional amniotic membrane graft (AMG) was done first for the right eye and a 360° peripheral lamellar corneal graft with additional AMG for the left eye. The BCVA of RE was 1/60 improving to 6/36 with plus ten aphakic glasses and LE was 3/60. Hand fashioned full thickness crescentic customised corneal graft with additional AMG and a peripheral 360° lamellar corneal graft with additional AMG in these cases are a novel approach to Mooren's ulcer with gratifying results.

Keywords: Mooren's ulcer, Bilateral, Perforation

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Introduction

Mooren's ulcer was first described by Bowman in 1849.[1] Mooren's ulcer is an idiopathic, rapidly progressive, painful, relentless, chronic ulcerative keratitis that begins peripherally and progresses circumferentially and centrally with no associated scleritis, and no detectable systemic disease. It is a diagnosis of exclusion which means all other diagnosable systemic disorders that could be responsible for the progressive destruction of cornea must be ruled out. The aetiology of Mooren's ulcer remains uncertain. However, recent studies indicate that it is an autoimmune disease directed against a specific target molecule in the corneal stroma, triggered in genetically susceptible individuals. A hand fashioned crescentic customised corneal graft is a well documented surgical approach for Mooren's ulcer but 360° peripheral lamellar corneal grafting is new approach.

Case report

A 60-year-old male presented to us with complaint of decreased vision, redness, watering, ocular pain and photophobia in both eyes for last 2 months more so in right eye for last one week. On examination, best corrected visual acuity in both eyes was counting finger close to face. Slit-lamp biomicroscopy of the Right eye showed inferotemporal crescent of corneal thinning involving 6 clock hours (5–11 o'clock) with limbal involvement but no associated scleritis (Fig. 1a) and there was a perforation with iris prolapse at 6–7 o'clock position. Right eye also showed a nuclear sclerotic cataract.

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Left eye showed 360° peripheral corneal thinning with melt and a central contact lens type cornea (Fig. 1b). In both eyes, the overhanging edges were oedematous and infiltrated with white cells which was spreading towards the centre and also circumferentially. It was positive for fluorescein stain indicating overlying epithelial defect. He was investigated to rule out systemic disease causing peripheral ulcerative keratitis such as Random blood sugar (RBS), Hemogram with sedimentation rate (ESR), fluorescent treponemal antibody absorption test (FTA-ABS), rapid plasma reagin (RPR), VDRL, angiotensin converting enzyme (ACE), anti-neutrophil cytoplasmic antibody (ANCA), antinuclear antibody (ANA), rheumatoid factor (RF), urine routine and microscopy, HCV, X-ray chest and Joints, SGPT, HBsAg. After excluding systemic diseases associated with peripheral ulcerative keratitis, a diagnosis of Bilateral Mooren's ulcer was made and systemic immunosuppressive therapy was started with oral cyclophosphamide 50 mg two times a day and oral systemic steroids at a dose of 1.5 mg/kg/day. Conjunctival resection was done in the area of melt. The dead tissue was debrided and the necrotic cornea was dissected (Fig. 2a and b) and cut carefully along the edges and the iris was pushed back and preserved. Lens was removed as it was intumuscent and bulging forward, no intra ocular lens was placed (Fig. 2c). A free hand fashioned full thickness cresentic customised corneal graft (Fig. 3a) with AMG (Fig. 3b) was done in the right eye. There was circumferential thinning which was progressing even with immunosuppressant's in the left eye, as there was a perforation in the right eye already. A week later in the left eye a 360° peripheral lamellar corneal graft along with AMG (Fig. 4a–c) was done as the ulceration was restricted to the periphery and the central part of the cornea was clear. The peripheral cornea was debrided and all the devitalised tissue was removed. A lamellar dissection was done, lamellar graft was taken and sutured 360° with partial thickness sutures. In the post-operative period, patient was

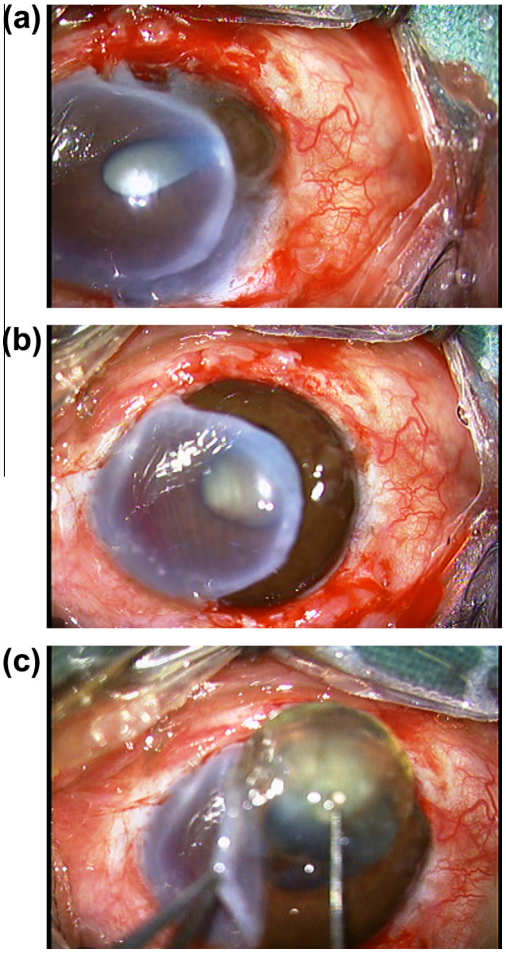

Figure 2. (a) Dead and necrotic area was debrided, (b) dead and necrotic area was removed, and (c) lens was removed.

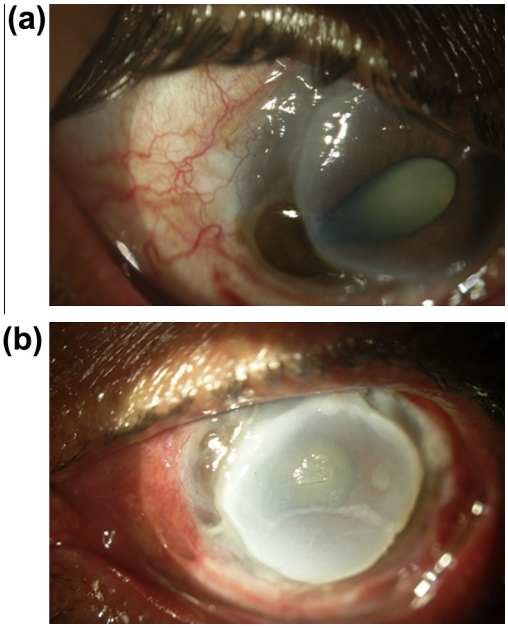

Figure 1. (a) Infero-temporal area of corneal thinning without scleritis, (b) left eye showed 360 peripheral corneal thinning.

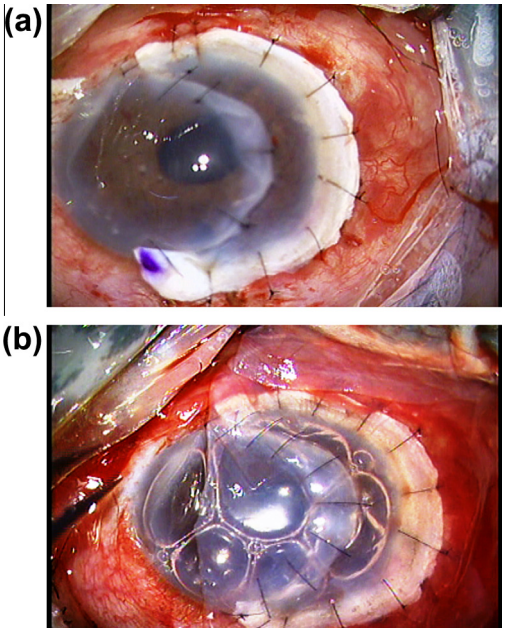

Figure 3. (a) Cresentic customised graft and (b) AMG placed over the graft.

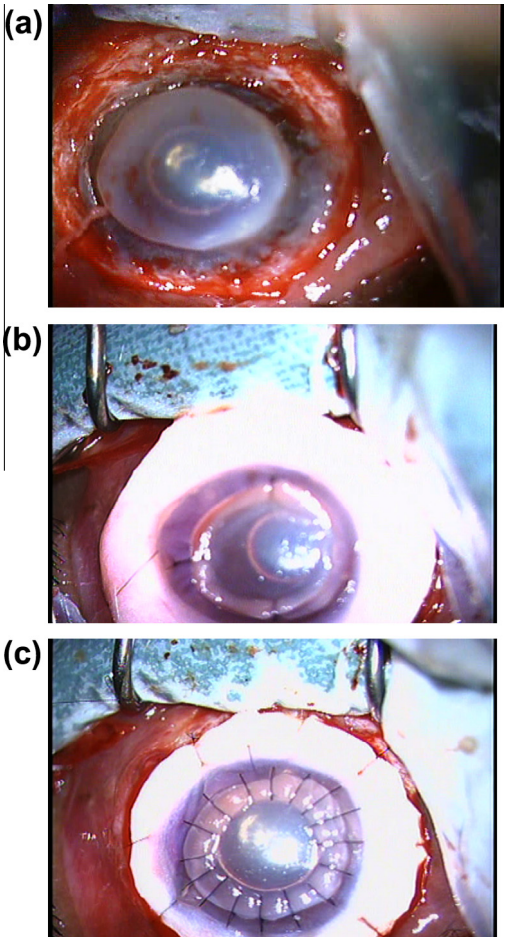

Figure 4. (a) Peripheral necrotic tissue was debrided, (b) peripheral lamellar graft, and (c) peripheral lamellar graft with AMG.

put on antibiotic steroid drops, homatropine eye drops and systemic steroids with 1 mg/kg body weight and cyclophosphamide was continued. The postoperative follow up at the end of 1 month, 3 months and 6 months showed graft had taken up well and no signs of recurrence of ulceration in both eyes.

Discussion

The diagnosis of Mooren's ulceration may be difficult when a patient first presents with PUK – the clinical appearance and its characteristic. However, a thorough medical history, physical examination is required. Mooren's ulcer is idiopathic by definition, occurring in complete absence of any diagnosable systematic disorder that could be responsible for progressive destruction of the cornea with no associated scleritis. Wood and Kaufman having reported 9 cases concluded that there were two clinical types of Mooren's ulcer.[2] The first limited type, is usually unilateral, with mild to moderate symptoms, generally responds well to medical and surgical treatment. This type is believed to occur in older patients and has become known as typical or benign Mooren's ulcer. In contrast, the second type is bilateral, with relatively more pain and generally a poor response to therapy in younger patients, became known as atypical or malignant, Mooren's ulcer. The benign type is bilateral in 25% of patients and the malignant type is bilateral in 75% of patients. Our case is different since it is bilateral in older age group. Lewallen and Courtright,[3] in their published series of Mooren's ulcer, suggest that younger patients had bilateral disease less frequently than older patients (1.5:1) regardless of race. They found that men were 1.6 times more likely to have Mooren's ulcer than women. It is extremely difficult to treat Mooren's ulcer and in many cases the results are poor. The treatment includes cortisone administered systemically and locally, but if this is unsuccessful, the complete excision of the perilimbal conjunctiva and episclera near the ulcer is made. Cyanoacrylate glue can be used in case of a small perforation. In the event of larger perforation, as in our case, the area can be covered with amniotic membrane or a lamellar keratoplasty can be performed. In this case, because of the presence of a large perforated corneal ulcer, a crescentic customised corneal graft with additional AMG was done. The percentage of surviving graft tissue has always been very low mainly because of the early onset of epithelial damage to the transplanted tissue, recurrence of the underlying disorder or secondary glaucoma caused by the disturbance of the iridocorneal angle and thereby of aqueous humour filtering.[4] The combination of corneal grafting and AMG helped as the area of perforation was covered and amniotic membrane helped in epithelisation and healing process. A crescentic customised graft with additional AMG in the right eye in our case is standard procedure with good results but our case report is different because instead of doing a total limbus – limbus full thickness graft for the left eye we did a 360° lamellar peripheral corneal graft thus salvaging the central part of the cornea which was not involved.

Conflict of interest

The authors declared that there is no conflict of interest.

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